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CASE REPORTS

Cerebral amyloid angiopathy presenting as a sequential cheiro-oral syndrome

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Abstract

Clinical recognition of cerebral amyloid angiopathy is important since aggressive antiplatelet or anticoagulation use may result in intracranial hemorrhage. Following case illustrates sequential evolving sensory disturbance involving perioral and ipsilateral finger (cheiro-oral syndrome) as a manifestation of underlying cerebral amyloid angiopathy.

Clinical recognition of cerebral amyloid angiopathy (CAA) remains difficult, since deposition of β-amyloid in vessel walls may manifest with varying clinical symptoms ranging from a non-specific, chronic occlusive process to a devastating, hyper-acute lobar hemorrhage. Recent clinico-radiologic studies suggest a subset of CAA cases, with contra-lateral convexial subarachnoid hemorrhage (cSAH) presenting with gradually evolving sensory disturbance lasting minutes ("amyloid march"), may be clinically recognizable in the elderly.

The following case illustrates sequential, bilateral sensory disturbance of homo-lateral finger and peri-oral tingling and numbness—cheiro-oral syndrome (COS)—as the predominant symptoms of CAA.

CASE REPORT

A 72-year-old retired marine officer, healthy without any vascular risk factors, presented with 3 episodes of right peri-oral and finger tingling and a "novocain" sensation over 2 days. Paresthesia would start at the angle of his mouth and finger tips, with a gradual spread to the cheek and up to his elbow over a 15 minute period. There was no confusion, headache or motor disturbance aside from a mild exacerbation of life-long familial tremor. He was not particularly alarmed since he recalled similar symptoms involving the opposite left peri-oral area and fingers 2 years previously. At that time the MRI scan reportedly revealed "small bleeding." A trial of oxcarbazepine seemed effective and was slowly tapered off one year later. This time, a brain CT on admission revealed chronic ischemic changes, most prominently on the right frontal lobe without obvious hemorrhagic foci; however, the MRI revealed an extensive multifocal focus of old hemorrhages concentrated near the gray-white matter junction, extending out into the cortical subarachnoid space, resulting in siderosis best seen on the gradient-echo (GRE) sequences (Figure 1). The CT angiography was unremarkable, and oxcarbazepine was restarted with symptomatic control. Further questioning of the patient revealed worsening memory disturbance, resulting in his transferring financial matters to his wife 2 months ago.

DISCUSSION

Detailed pathologic studies in CAA have revealed early, predominant, cortical, meningeal artery deposition of β-amyloid on the vessel wall leading not only to occlusive vasculopathy but also to hemorrhages from underlying fibrinoid necrosis, intimal fragmentation, microaneurysm formation and reactive inflammation. An adjacent subarachnoid hemorrhage is almost always found.[1] Even the well-known catastrophic lobar hemorrhage is likely caused from a large meningeal artery rupture in the depths of the sulci, with bleeding extending into the parenchyma.[2]

Recent case studies supplemented by pathologic confirmation suggest a subgroup of CAA cases presenting with non-traumatic, convexial subarachnoid hemorrhage (cSAH) maybe clinically recognizable.[3,4] When symptomatic, an elderly patient will present with evolving transient ischemic attack (TIA)-like symptoms, but with a gradual spread of typically positive,

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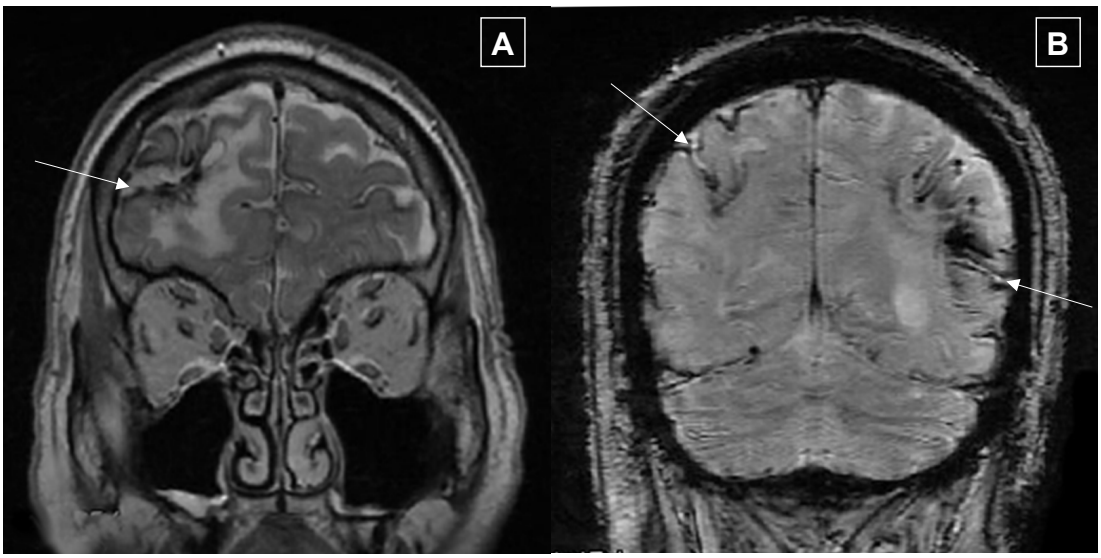

Figure 1 A, B. Coronal T2-weighted GRE sequences reveal areas of focal siderosis (arrows).

sensory phenomenon involving the homo-lateral limbs or the face over a span of several minutes. These migraine-aura-like episodes, nearly always without a headache, may demonstrate a contra-lateral focal cSAH on a CT scan. Furthermore, on a GRE or susceptibility-weighted image (SWI) MR sequences, a majority will reveal previously asymptomatic, cortical microhemorrhage or siderosis attributable to chronic oozing of blood into the subarachnoid space that is usually adjacent to the central sulcus.[3] Indeed, the presence of these characteristic lesions in 0.7% of an asymptomatic elderly population group further support the view that these changes represent an early manifestation of CAA.[5] Other causes of non-traumatic cSAH, such as cortical vein thrombosis, aneurysmal or arteriovenous malformation rupture, and vasculitis can also be considered but unlikely to present with sequential COS.

COS, a sensory disturbance restricted to the homo-lateral fingers and hand and peri-oral area, is usually indicative of underlying ischemia involving the descending tract from the contralateral sensory cortex to the caudal brainstem. Sequential and paroxysmal COS as the main clinical feature, as discussed here, are consistent with blood-brain-barrier leakage and vascular dysautoregulation involving the outer sensory cortical surface from a non-ischemic etiology, as previously reported in cortical arteriovenous malformation and subdural hematoma.[6]

Recognition of these TIA-like episodes as a clue to the presence of a contra-lateral cSAH, which may not be apparent on the brain CT, has important clinical implications. In the era of evolving clinical indication of IV-tPA use, even in cases of rapidly resolving stroke or a TIA due to demonstrated low bleeding risk with potential benefit[7], significance of this "amyloid march" should not be dismissed. Similarly, in the setting of chronic atrial fibrillation in the elderly, these "TIAs" should not be construed as an ischemic cardio-embolic phenomenon. Aggressive use of thrombolytic or antiplatelet agents may be risky.

DISCLOSURE

Conflict of interest: None

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